The Anomalous Origin of the Right Main Bronchus from the Esophagus *

JOHN L. KEELEY, M.D., ARNE E. SCHAIRER, M.D.

From the Departments of Surgery, Stritch School of Medicine of Loyola University and South Shore Hospital, Chicago, Illinois

SOME unusual problems were recently encountered during the repair of esophageal atresia and tracheo-esophageal fistula in a four day old infant. Postmortem examination revealed the anomalous origin of the right main bronchus from the esophagus.

Although there is great variety in the anomalies associated with esophageal atresia and tracheo-esophageal fistula we have not found this particular anomaly previously reported in the available literature.

Case Report

Shortly after birth a five pound six and one-half ounce male infant was cyanotic, and frequent aspiration of mucus from the oropharynx was necessary. A diagnosis of hyaline membrane disease was made and feedings were withheld. On the third day x-ray examination with Lipiodol® demonstrated esophageal atresia (Fig. 1). The film also showed a gastro-intestinal gas pattern which proved the presence of a tracheo-esophageal fistula involving the lower esophageal segment. The blind esophageal pouch terminated at the level of the thoracic inlet. The trachea and the left main bronchus were outlined by some of the contrast media which was thought to have been aspirated after "spilling over" from the esophageal pouch. Two opaque lines were noted near the midline and in retrospect suggest the outline of the distal esophageal segment. Failure to demonstrate the right main bronchus suggested obstruction due to mucus. Haziness in the right upper lung field was compatible with pneumonitis or atelectasis. A 12-hour period of preparation improved his poor general condition, and on the fourth day operation was performed.

General anesthesia was induced with open drop ether. The endotracheal tube was readily introduced into the larynx, but resistance which obstructed the endotracheal tube was encountered a few millimeters beyond. By slightly withdrawing the endotracheal tube, maximal ventilation was attained. The infant was placed on his left side and the chest was opened through the right, fourth interspace. The right lung had no fissures. It was partially aerated, but in contrast to the left lung, showed no ventilatory excursions. When expanded by increased pressure on the anesthetic bag, it remained distended for several minutes, then gradually became atelectatic. This series of events was observed three or four times.

The blind pouch and the tracheo-esophageal fistula were found at the level of the thoracic inlet, an unusually high position for either of these structures. Gas escaped from the lower esophageal segment as the fistula was divided. The tracheal side of the fistula was closed with fine black silk sutures. At this time the right lung became atelectatic, and remained so despite further attempts to expand it by pressure applied through either the endotracheal tube, which later became dislodged, or thereafter through a tight-fitting face mask. Repeated tracheal aspirations to remove a "bronchial plug" were likewise unsuccessful.

A two-layer end-to-end anastomosis of the esophageal segments over a small catheter was accomplished quite readily. The condition of the patient demanded termination of the operation despite the inability to inflate the right lung. The chest wound was closed in layers and the pleural space was drained with a fine catheter attached to a water seal drainage system. The baby was placed in a warm humid environment immediately and, despite high oxygen concentration, frequent attacks of cyanosis were noted. Six hours after the operation the infant expired.

The autopsy disclosed a sound anastomosis. The right lung was completely atelectatic as was much of the left lung. Neither had fissures. Dissection of the mediastinal structures demonstrated the following interesting and revealing findings (Fig. 2A, B). Two cm. distal to the vocal cords the trachea narrowed sharply from 3.5 to 1.5 mm. in diameter. This narrow trachea continued for 47 mm. where it was connected with the hilum of the left lung. The right main bron-

* Submitted for publication February 1, 1960.

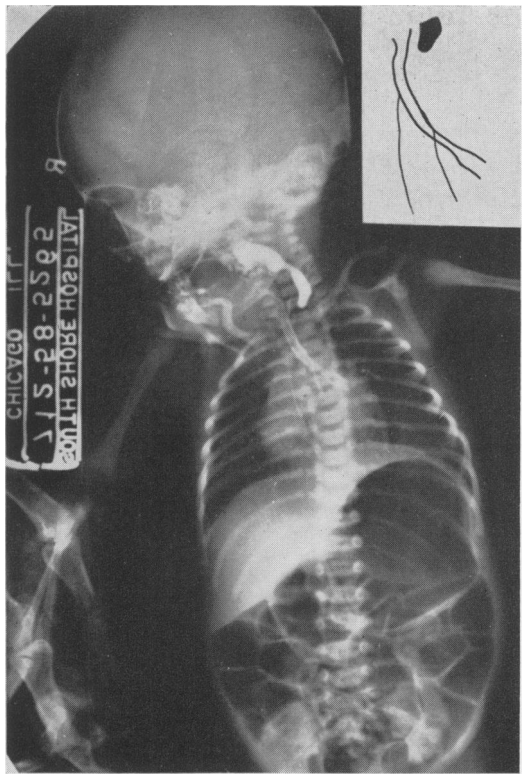

FIG. 1. A roentgenogram showing Lipiodol® in the proximal esophageal pouch. Some has "spilled over" into the trachea outlining the left main bronchus. Two fine lines faintly outline the distal esophageal pouch. The insert shows a tracing of these shadows.

chus was 10 mm. long and 1.5 mm. in diameter. Its lumen communicated with that of the esophagus without constriction at a point 22 mm. above the cardio-esophageal junction. Microscopic examination of the esophagus and bronchus revealed normal histological findings for each structure. The actual junction of the stratified squamous epithelium of the esophagus with the ciliated pseudo-stratified columnar epithelium of the bronchus, however, is not available for study. Except for atelectasis and areas of pneumonitis the histology of the lungs was normal.

The abnormalities found at autopsy fully explain a number of events. The narrowing of the trachea accounts for the difficulty with the endotracheal intubation. The tube became obstructed when it encountered the narrow portion. With the arrangement as shown in Figure 2 aeration of the right lung could be accomplished only by air passing into the trachea, through the fistula, into the lower esophageal segment and then into the right main bronchus. Once the fistula was divided, this route was interrupted.

Upon the completion of the esophageal anastomosis, a new route for the entrance of air into the right bronchus was established. However, this pathway was obstructed during the performance of the anastomosis due to the catheter over which the anastomosis was made. After the catheter was withdrawn, this avenue for entry of air into the right bronchus proved to be ineffective as the atelectasis of the right lung persisted.

Discussion

A resumé of the embryology of the normal esophagus and respiratory apparatus provides only a background for speculation to explain the origin of a bronchus from the esophagus.[1] The respiratory apparatus and the esophagus both arise from the foregut. The lung buds appear first as an outpouching on the ventral surface of the foregut. Soon this outpouching splits into two lung buds. A less prominent protrusion

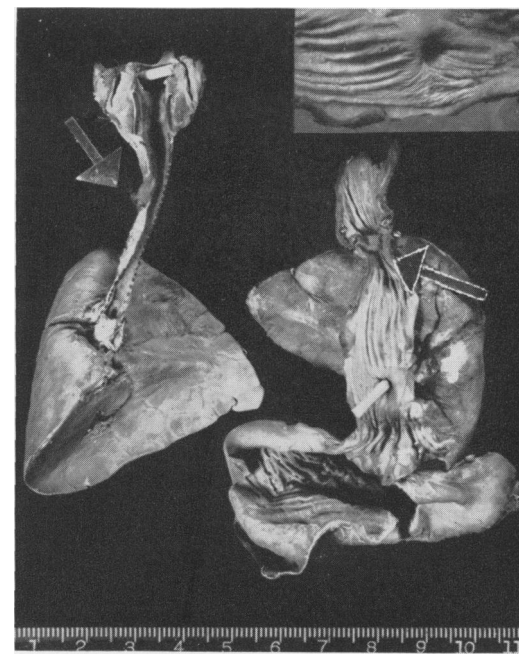

FIG. 2A. A posterior view of the specimen showing the origin of the right main bronchus from the esophagus. The arrow on the left indicates the site of the closed fistula. The arrow on the right identifies the site of the esophageal anastomosis. An applicator stick has been passed into the right main bronchus and protrudes through the bronchial orifice in the esophagus. The insert shows the folds of esophageal mucosa surrounding the bronchial orifice.

from the foregut appears cranial to the lung buds and extends cephalad. This later separates from the foregut and becomes the trachea. The separation of these two hollow structures proceeds in the cranial direction. The larynx develops at the point where this separation normally ceases.

The theories of pathogenesis of tracheo-esophageal fistulas are numerous. The effect of the pressure from adjacent vascular anomalies has been suggested as a cause; however, it seems unlikely. In a report of 224 patients operated upon by Gross [7] for esophageal atresia no mention is made of any problem due to pressure from abnormalities of the great vessels. Moreover, in his series of 57 patients seen with enough compression due to abnormalities of the great vessels to require surgical correction, there was no instance of esophageal atresia or tracheo-esophageal fistula.

A careful study of the literature reveals only two other patients with a bronchial connection to the esophagus. Gans and Potts [6] report a five-month-old patient who had fever and cough and who failed to gain weight. Investigation revealed an opacity in the left upper lung field. During bronchoscopy the left upper lobe bronchial orifice appeared to be compressed and an attempt to pass a flexible curved aspirator into this bronchus was unsuccessful. Esophagoscopy disclosed the presence of an orifice, apparently lined with normal mucous membrane, in the wall of the esophagus at the level of the bifurcation of the trachea. Rather thick pus was seen to escape from this opening with each respiration. Bronchograms demonstrated a depressed left bronchus and no Lipiodol® entered the left upper lobe bronchus.

Thoracotomy was performed and a soft mass about 8 cm. in diameter was removed from the left upper chest. A fistula to the esophagus was indentified, divided and sutured. Although the mass was adherent to the left main bronchus, it did not communicate with it. The pathologic diagnosis

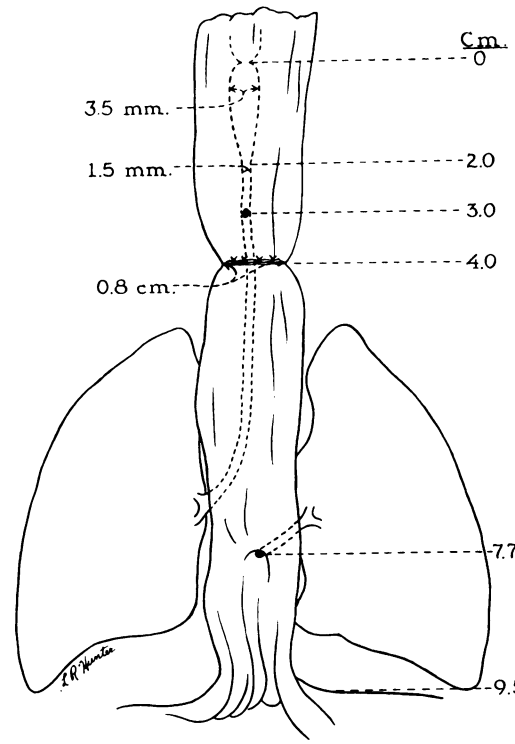

FIG. 2B. This drawing illustrates the findings at autopsy as viewed from the posterior. The narrowed trachea, the right main bronchus arising from the esophagus, the esophageal anastomosis, and the closed site of the fistula are shown together with the significant dimensions.

was ectopic bronchus originating from the esophagus and lung tissue with chronic aspiration pneumonia. This patient's lobe differed from the usual anomalous lobe by apparently representing or replacing the normal left upper lobe. It received blood from two branches of the pulmonary artery.

The second instance of this anomaly has been reported by St. Raymond, Hardy and Robbins [9] whose 13-year-old patient had intermittent attacks of cramping epigastric pain. A roentgenogram showed a translucent area in the left lower chest. Contrast studies of the gastrointestinal tract revealed displacement of the stomach into the chest, confirming the diagnosis of hiatus hernia. A left thoracotomy to repair the hernia disclosed a bluish-gray mass attached by a pedicle to the esophagus. While dissecting the mass from the esophagus it

was found to communicate with it just above the cardio-esophageal junction. The pathologic diagnosis of the excised specimen was accessory lobe of the lung showing chronic interstitial inflammation.

There are numerous reports of the pulmonary anomaly referred to as lower accessory lung, supernumerary lung or Rokitansky's lobe. In 1944, Davies and Gunz [3] reported two patients with "lower accessory lung." They found "about forty" such cases in their review of the literature but did not include two which had been reported by Freedlander and Gebauer, in 1939. Two additional instances have been reported, one by DeBakey, Arey and Brunazzi,[4] and the other by Leahy and MacCallum.[8] Carter and Osborn [2] have suggested the term aberrant rather than accessory or supernumerary lung to differentiate these anomalies from accessory lobes which have bronchial communications.

This "aberrant" lung tissue is usually located in the left hemithorax just above the diaphragm. However, it may be found in either side of the chest, in the lower neck or upper abdomen, and a diaphragmatic hernia is frequently associated with it. It is not connected with the normal respiratory tract by a bronchus and its blood supply, with rare exceptions, is systemic in origin rather than from the pulmonary artery.

A review of the description of the specimens that have been reported as accessory lungs discloses an extremely variable picture. In some instances the lung tissue is cystic, and microscopic examination reveals atypical pulmonary histology. Despite the description of wide variations in these lesions and their bizarre anatomic relations, careful study disclosed only the two instances of communication between a lobar bronchus and the esophagus described above. We were unable to find any report of a main bronchus arising from the esophagus.

Summary

A patient with esophageal atresia and a tracheo-esophageal fistula with origin of the right main bronchus from the esophagus is reported. Operation on the fourth day of life was complicated by inability to expand the right lung. The patient died in the early postoperative period. Postmortem examination revealed that the right main bronchus arose from the esophagus, which explains the problems in expanding the right lung at the operation.

The differences between "anomalous pulmonary tissue" noted frequently in the literature and the anomalies found in our patient are emphasized. This appears to be the first report of a main bronchus arising from the esophagus.

Bibliography

1. Arey, L. B.: Developmental Anatomy, 4th Ed., Philadelphia, W. B. Saunders Co., 1942, p. 231.
2. Carter, H. and H. A. Osborn: Accessory Lobe of Lung. J. Obst. & Gynaec. Brt. Emp., **43**: 1194, 1936.
3. Davies, D. V. and F. W. Gunz: Two Cases of Lower Accessory Lung in the Human Subject. J. Path. & Bact., **56**:417, 1944.
4. DeBakey, M., J. B. Arey and R. Brunazzi: Successful Removal of Lower Accessory Lung. J. Thoracic Surg., **19**:304, 1950.
5. Freedlander, S. W. and P. W. Gebauer: Diseases of Aberrant Intrathoracic Lung Tissue. J. Thoracic Surg., **8**:581, 1939.
6. Gans, S. L. and W. J. Potts: Anomalous Lobe of Lung Arising from the Esophagus. J. Thoracic Surg., **21**:313, 1951.
7. Gross, R. E.: Surgery of Infancy and Childhood. Philadelphia, W. B. Saunders Co., 1953, p. 95 and p. 913.
8. Leahy, L. J. and J. D. MacCallum: Cystic Accessory Lobe. J. Thoracic Surg., **20**:72, 1950.
9. St. Raymond, Jr., A. H., J. D. Hardy and S. G. Robbins: Lower Accessory Lung Communicating with the Esophagus and Associated with Congenital Diaphragmatic Hernia. J. Thoracic Surg., **31**:354, 1956.